

Durham Research Online

Deposited in DRO:

21 October 2016

Version of attached file:

Accepted Version

Peer-review status of attached file:

Peer-reviewed

Citation for published item:

Lough, E. and Flynn, E. and Riby, D. M. (2016) 'Personal space regulation in Williams syndrome : the effect of familiarity.', *Journal of autism and developmental disorders.*, 46 (10). 3207-3215.

Further information on publisher's website:

<http://dx.doi.org/10.1007/s10803-016-2864-8>

Publisher's copyright statement:

The final publication is available at Springer via <https://doi.org/10.1007/s10803-016-2864-8>

Additional information:

Use policy

The full-text may be used and/or reproduced, and given to third parties in any format or medium, without prior permission or charge, for personal research or study, educational, or not-for-profit purposes provided that:

- a full bibliographic reference is made to the original source
- a [link](#) is made to the metadata record in DRO
- the full-text is not changed in any way

The full-text must not be sold in any format or medium without the formal permission of the copyright holders.

Please consult the [full DRO policy](#) for further details.

Abstract

Personal space refers to a protective barrier that we strive to maintain around our body. We examined personal space regulation in young people with Williams syndrome (WS) and their typically developing, chronological age-matched peers using a parent report questionnaire and a stop-distance paradigm. Individuals with WS were reported by their parents to be more likely to violate the personal space of others, and indeed they maintained a shorter interpersonal distance in the stop-distance paradigm. Interestingly, WS individuals failed to regulate their personal space based on the familiarity of the person they were interacting with. Findings are discussed in relation to the wider social profile associated with WS, and the possible impact of atypical personal space regulation on social vulnerability.

Personal space regulation in Williams syndrome: The effect of familiarity

When engaging in social interactions, individuals must regulate the distance that they maintain between themselves and other people (Hall, 1996). Personal space is defined as the area around a person's body, which if invaded, can cause feelings of discomfort and anxiety (Perry et al., 2013). Vignemont and Iannetti (2015) outline different types of personal space. Peripersonal space refers to the space around one's body where an object can be grasped, whereas extrapersonal space is the area around the body that is just beyond reach. Indeed, they highlight that there also exists functional differences within the definition of peripersonal space. The Function-Specific Model of personal space identifies two functions of peripersonal space: the protective (or defensive) space (Sambo & Iannetti, 2012) and the working personal space (Rizzolatti et al., 1997). The current study is concerned with the protective peripersonal space (herein referred to as simply 'personal space'), and its implications for social vulnerability levels in Williams syndrome.

For typically developing individuals, regulating this personal space is a largely automatic process, guided by situational cues, social cues and cultural norms (Beaulieu, 2004). The ability to successfully collate these cues and maintain an appropriate interpersonal distance contributes to successful and positive social interactions (Gessaroli et al., 2013). However, it is known that some individuals with developmental disorders find social interactions challenging, and they may also struggle to regulate their personal space. Gessaroli et al. (2013) studied personal space regulation in children with autism spectrum disorders (n=15, mean 9 years; ASD) and compared them to typically developing (n=23, mean 9 years; TD) children. Using a stop-distance paradigm, they found that children with ASD maintained a greater distance from a confederate compared to their TD peers. Further, whilst TD children were able to regulate their personal space based on the familiarity of the

person they were interacting with, children with ASD failed to do so, suggesting that they lack flexibility in personal space regulation.

Kennedy and Adolphs (2014) also examined the issue of personal space in young people with ASD. They used the Social Responsiveness Scale (SRS; Constantino & Gruber, 2005), which is a 65 item parent report questionnaire designed to measure the typicality/atypicality of social functioning. They were specifically interested in item 55 (“Knows when he or she is too close to someone or is invading someone’s space”). In stark contrast to the findings of Gessaroli and colleagues (2013), Kennedy and Adolphs (2014) found that individuals with ASD were more likely to be reported by parents to violate the personal space of others. Indeed, 79% of parents report that their children with ASD have smaller personal space boundaries compared to their TD siblings. However, the different methods of assessment used in these studies do not allow for direct comparisons of results, and if personal space regulation is disorder-specific, then they offer little insight in to how individuals with other developmental disorders regulate their personal space.

Williams syndrome (WS) is a rare neuro-developmental disorder, which affects approximately 1 in 20,000 individuals (Korenberg et al., 2003). It is caused by the microdeletion of 25-28 genes on chromosome 7 (7q11.23; Hiller et al., 2003). Individuals with WS typically have mild-moderate levels of intellectual impairment (Searcy et al., 2004), and experience a powerful pro-social drive to interact with others, i.e., they display a hypersocial behavioural phenotype (Jarvinen et al., 2013). Despite their social nature, individuals with WS can struggle to pick up on social cues, and many find it hard to form and maintain peer relationships, resulting in high levels of isolation (Udwin, 1990). This occurs against a backdrop of high anxiety levels (Stinton et al., 2010). Recent work by Riby and colleagues (2014) found that 46% of children and adults with WS experienced high levels of anxiety, with a mean for this high anxiety group above that found in clinically anxious

children (Nauta et al. 2004). Interestingly, they also noted differing patterns of social behaviour, as measured by the Social Responsiveness Scale (SRS). Those individuals who experience higher anxiety showed more severe social dysfunction, suggesting that anxiety levels are linked to social behaviour in WS. As individuals with WS show indiscriminate approach behaviour (Little et al., 2013), and a lack of stranger danger awareness (Riby et al., 2013), their personal space regulation when interacting with others is an important facet when looking at their social vulnerability profile (Jawaid et al., 2012; Lough et al., 2015b).

Lough et al. (2015a) offered the first insights in to personal space regulation in WS and ASD, using the same methods employed by Kennedy and Adolphs (2014). They found that individuals with WS were reported by their parents to show the least awareness of personal space boundaries, when compared to reports from parents of individuals with ASD and TD individuals. This is of particular concern given the wider social vulnerability profile associated with WS (see Jawaid et al., 2012 for a review). Despite the large sample size used by Lough and colleagues, it was acknowledged that the SRS was not designed to measure personal space, and using a method such as the stop-distance paradigm could provide more clarity and insight in to this issue. In the current study, we used both the SRS and the stop-distance paradigm to provide multiple measures of personal space regulation in young people with Williams syndrome. Our aim was to obtain a more comprehensive insight in to this issue by utilising information from a parent report questionnaire and experimental work involving the individuals with WS themselves. Based on the work of Lough et al. (2015a), as well as what we know about the WS social profile, large effect sizes were predicted, and the hypotheses were as follows: 1) Children and adolescents with WS would receive higher scores on SRS items relating to personal space than their TD peers; 2) In the stop-distance task, young people with WS would let unfamiliar people approach and stand at a closer

interpersonal distance than TD children and adolescents, and 3) In the same task, young people with WS would approach and stand closer to an unfamiliar person than their TD peers.

Method

Participants

Eighteen young people with WS (mean age = 11.4; age range = 8 – 16) and eighteen typically developing children and adolescents (mean age = 11.3; age range = 8 – 16) participated in the study (see Table 1). An a priori power analysis indicated that 12 participants were needed in each group to have 80% power for detecting a large effect size ($d = 0.8$, $\alpha = 0.05$). The participants were matched on chronological age and gender. All participants with WS had previously had their diagnosis confirmed using *fluorescent in situ hybridization* (FISH) testing.

Social Responsiveness Scale (Constantinno & Gruber, 2005)

This parent report questionnaire consists of 65 items which measure the typicality/atypicality of social functioning. It has frequently been used in the typically-developing population, but also with young people with WS (e.g. Klein-Tasman et al., 2011; Riby et al., 2014; Lough et al., 2015a). From the responses, five sub-scale scores can be generated in the areas of: social awareness, social cognition, social communication, social motivation and autistic mannerisms (see Table 1). Higher scores suggest greater levels of impairment.

There are several items relating to personal space that have been examined in the work of Kennedy and Adolphs (2014) and Lough and colleagues (2015a) and are therefore of interest to the current study. They examined item 55 which asks parents to rate the following statement: “knows when he or she is too close to someone or is invading someone’s space”.

Other items also examined by both Kennedy and Adolphs (2014) and Lough et al., (2015a) refer to multi-modal construct of personal space, including item 52 (“Knows when he or she is talking too loud or making too much noise”), item 56 (“Walks in between two people who are talking”) and item 63 (“Touches others in an unusual way e.g. he or she may touch someone just to make contact with them then walk away without saying anything”).

Spence Children’s Anxiety Scale – Parent version (Spence, 1998)

The Spence Children’s Anxiety Scale–Parent Version (SCAS-P; Spence, 1998) was used to assess symptoms of anxiety. This issue is particularly relevant due to increased anxiety in WS and reports of an association between increased anxiety and atypical social behaviours (Riby et al., 2014). The SCAS-P has been reported to have good psychometric qualities including a high internal consistency of .92 (Spence, Barrett, & Turner, 2003), and has been used in both TD and clinically anxious populations (Nauta et al., 2004; Spence, 1998). This 38-item parent report questionnaire is divided into six subscales of anxiety relating to panic/agoraphobia, separation anxiety, physical injury fears, social phobia, obsessive compulsive, and generalised anxiety disorder. Parents rate each item on a 4-point Likert scale according to how often their child exhibits the symptoms, from 1 (never) to 4 (always). Their answers are scored from 0 to 3, yielding a maximum possible score of 114. While there is no formal clinical cut-off for the SCAS-P, total SCAS scores of 24 or above have been suggested to indicate clinical levels of anxiety (Spence, 2008).

[Table 1]

Procedure

A stop-distance paradigm was used to assess preferred interpersonal distance (Kennedy et al., 2009). This procedure has been used extensively for assessing preferred

interpersonal distance under different conditions, yielding reports of high reliability and validity (Greenberg, Strube, & Myers, 1980; Hayduk, 1978, 1983, 1985). The task began with the participant standing 3 metres away from the experimenter. There were four conditions: two of which involved completing the task with an unfamiliar person (the experimenter) and the other two were undertaken with a familiar person (mother/father; see Figure 1). In condition A the participant was asked to approach the experimenter and stop at a location that felt comfortable to them. The experimenter maintained a neutral expression and no eye contact was made. Once the participant had decided on a location which felt comfortable to them, a hip-to-hip measurement was taken using a digital laser distance measurer (RZE-40). Three measurements were taken in succession and averaged together. Each condition consisted of three trials. The average distance across these three trials was taken as the preferred distance in each condition. This procedure was repeated for condition B, in which the experimenter approached the participant and the participant instructed them to stop at a distance that felt comfortable. Conditions C and D mirrored the first two conditions, but a familiar person took the place of the experimenter. The conditions were presented in a random order for each participant. Ethical approval was obtained from the host institution.

[Figure 1]

Results

Social Responsiveness Scale

To examine the overall social profile of the two groups, Mann-Whitney U tests were used. We found statistically significant differences between groups on the SRS total T scores, as well as on all of the five subscales (all $p < 0.01$). In all cases the WS group showed more atypical social behaviour. 92% of TD young people displayed overall social behaviour that was deemed to be in the normal range of functioning, whereas only 18% of young people with WS scored within the normal range (this maps directly onto levels reported in van der Fluit et al., 2012 and Riby et al., 2014). Importantly, the results also revealed statistically significant differences between the two groups on the four items in the SRS that relate to personal space, with parents of individuals with WS reporting greater atypicalities in their son/daughter on these items (all $p < 0.01$; Mann-Whitney U tests; see Figure 2).

[Figure 2]

Stop-distance paradigm

Being Approached by an Adult

A two-way mixed methods ANOVA was used on the measurement of interpersonal distance (m), with Familiarity (unfamiliar, familiar) as a within-subjects variable and Group (TD, WS) as a between subjects variable. There was a significant main effect of Familiarity ($F(1,34) = 4.74$, $p < 0.05$; $\eta_p^2 = 0.12$), showing that participants maintained a larger distance when approached by an unfamiliar person compared to a familiar person. There was also a significant main effect of Group ($F(1,34) = 4.75$, $p < 0.05$; $\eta_p^2 = 0.12$), as young people with WS showed reduced interpersonal space compared to their TD peers when they are being approached. Crucially, there was a significant interaction between Familiarity and Group

($F(1,34) = 15.18, p < 0.001; \eta_p^2 = 0.31$; see Figure 3), showing that individuals with WS maintained a much closer distance when approached by unfamiliar people than TD individuals do, with a large effect size. Post hoc analyses showed that the WS group ($M = 0.74, SD = 0.29$) and the TD group ($M = 0.75, SD = 0.2$) did not significantly differ in their interpersonal distance when approached by a familiar person ($t(34) = -0.13, p = 0.9, d = 0.04$; independent t-test). However, when approached by an unfamiliar person, the WS group ($M = 0.68, SD = 0.26$) let the unfamiliar person stand at a significantly closer distance to them than the TD group ($M = 0.98, SD = 0.22; t(34) = -3.7, p < 0.001, d = 1.3$). Indeed, there was a significant difference for the preferred interpersonal distance of the TD group when approached by familiar versus unfamiliar people ($t(17) = 4.8, p < 0.001$), but this was no significant difference for the WS group ($t(17) = 1.28, p = 0.22$).

Approaching an Adult

The above tests were repeated in order to examine interpersonal distance when the child was doing the approaching. A two-way mixed methods ANOVA on Familiarity (unfamiliar, familiar) and Group (TD, WS) showed a significant main effect of familiarity ($F(1,34) = 15.6, p < 0.001; \eta_p^2 = 0.31$). When the child is the one approaching, there was no significant main effect of Group ($F(1,34) = 2.71, p = 0.11; \eta_p^2 = 0.07$). However, there was a large effect size and a trend towards a significant interaction between Familiarity and Group ($F(1,34) = 3.72, p = 0.06; \eta_p^2 = 0.1$; see Figure 3), with the WS group coming much closer when they approach unfamiliar people compared to the TD group. Between subjects *t*-tests revealed that there was no difference between groups when approaching a familiar person (WS: $M = 0.7, SD = 0.23$; TD: $M = 0.76, SD = 0.22; t(34) = -0.77, p = 0.44$). There was, however, a significant difference between groups when approaching an unfamiliar person, with the TD group standing further away ($M = 0.97, SD = 0.31$) than the WS group ($M = 0.78, SD = 0.25, t(34) = -2.08, p < 0.05, d = 0.7$). The TD individuals showed a significant

difference between the distance they maintained when approaching a familiar adult compared to an unfamiliar adult ($t(17) = 6.1, p < 0.001$), whereas no significant difference was found for the WS group ($t(17) = -0.99, p = 0.34$).

[Figure 3]

Factors impacting on personal space regulation

Age

In the WS group, there was no significant correlation between age and the distance these individuals maintained between themselves and the experimenter ($r = 0.28, p = 0.27$). However, there was a correlation between age and how close they let a stranger come to them, with older individuals requesting that the unfamiliar person kept a greater distance ($r = 0.53, p < 0.05$). There was no correlation between age and interpersonal distance found in any of the conditions for TD individuals

When looking at age effects on item 55 of the SRS, there was a significant negative correlation between age and the likelihood of violating another person's personal space in the WS group ($r = -0.62, p = 0.01$). No significant correlation was found between age and item 55 for the TD group ($r = -0.08, p = 0.75$).

Anxiety

There was a significant difference between the WS group ($M = 26.78, SD = 14.38$) and the TD group ($M = 14.56, SD = 6.32; t(34) = 3.3, p < 0.01, d = 1.1$) on their total SCAS scores, with higher anxiety in the WS group. There was no significant correlation between anxiety (SCAS total scores) and the distance maintained when approaching a familiar (WS: $r = -0.29, p = 0.24$; TD: $r = -0.25, p = 0.32$) or unfamiliar person (WS: $r = -0.22, p = 0.07$; TD: $r = -0.43, p = 0.07$). Anxiety did not correlate with approach by a familiar person (WS: $r = -$

0.62, $p = 0.81$; TD: $r = -0.21$, $p = 0.39$), but there was a significant negative correlation between anxiety and how close both groups let an unfamiliar person stand from them (WS: $r = -0.55$, $p < 0.05$; TD: $r = -0.52$, $p < 0.05$), suggesting that young people high in anxiety let unfamiliar people stand closer to them. Although this might appear counterintuitive this result requires further investigation as it may be that there is a bi-directional effect where anxiety also feeds off an inability to make appropriate judgements about others in such social situations.

Relationship between scores on the Social Responsiveness Scale and stop-distance paradigm

There was no significant correlation between the four conditions and the individuals' total SRS t scores in the WS group (Condition A: $r = -0.04$, $p = 0.87$; Condition B: $r = -0.38$, $p = 0.12$; Condition C: $r = -0.24$, $p = 0.33$; Condition D: $r = -0.02$, $p = 0.94$), or in the TD group (Condition A: $r = -0.14$, $p = 0.59$; Condition B: $r = -0.02$, $p = 0.95$; Condition C: $r = -0.03$, $p = 0.91$; Condition D: $r = -0.09$, $p = 0.71$). Similarly, overall approach behaviour was not significantly correlated with the SRS items related to personal space in the WS group (Item 52: $r = 0.12$, $p = 0.64$; Item 55: $r = -0.16$, $p = 0.53$; Item 56: $r = -0.19$, $p = 0.44$; Item 63: $r = -0.34$, $p = 0.17$) or the TD group (Item 52: $r = -0.21$, $p = 0.39$; Item 55: $r = -0.26$, $p = 0.3$; Item 56: $r = -0.1$, $p = 0.69$; Item, 63: $r = -0.12$, $p = 0.63$). Of interest, there was a significant negative correlation found between being approached by an unfamiliar person (Condition B) and levels of social awareness on the SRS ($r = -0.54$, $p < 0.05$); i.e., those with the most impaired levels of social awareness let unfamiliar people come closer to them.

Discussion

Individuals with WS are reported by their parents to be more likely to violate the personal space of others than their TD peers, supporting our first hypothesis. This replicates the findings by Lough et al (2015a); reinforcing the notion that interpersonal distance regulation is highly atypical in individuals with WS, although the participant demographics between the two groups differ. In the second part of the study, the stop-distance paradigm was utilised, and found that individuals with WS maintained an overall shorter interpersonal distance than the TD individuals. Indeed, differences between the young people with WS and their TD peers were found only in their interpersonal distance around unfamiliar people, not familiar people. Specifically, young people with WS maintained a smaller interpersonal distance when approaching, and when being approached by, unfamiliar people, supporting the second and third hypotheses. Taken together with the findings from the SRS, it would seem that young people with WS show atypical interpersonal distance behaviour, and struggle to regulate their personal space in accordance with the familiarity of the person with whom they are interacting. This is the first study to empirically study social distance violations in this population and shows that this is a critical issue in Williams syndrome.

Our current findings also highlighted a significant negative correlation between anxiety and how close both individuals with WS and TD individuals let an unfamiliar person stand from them. This finding requires further investigation. It may be particularly relevant here to examine the role of social cognition in personal space regulation or vice versa. Individuals with WS have been shown to have deficits in understanding and predicting the actions of others (Sparaci et al., 2012). They show more pronounced difficulties in understanding ‘what’ action is being performed than individuals with ASD, and when compared to TD individuals matched on chronological and mental age (Sparaci et al., 2014). Both individuals with WS and those with ASD also showed impaired ‘why’ understanding. This may be pertinent in the context of the stop-distance paradigm, in which an approaching

act is being performed, and participants are required to read the observed actions in order to respond appropriately. Indeed, Tager-Flusberg and Sullivan (2000) proposed that there exists dissociation in WS between perceptual and cognitive components of social intelligence. They argue that whilst many individuals with WS are able to make immediate judgements about the mental states of other people (perceptual judgements), they struggle to make inferences about the content of these mental states (cognitive judgements). Elements of social cognition are likely to have a direct bearing on social approach behaviour in WS, and therefore require further investigation.

These findings are of particular concern given what is known about the indiscriminate approach behaviour using face-rating tasks with WS adults (Jones et al., 2000) and the lack of stranger danger awareness in WS (Riby et al., 2014). Invading the personal space of others, particularly strangers, can also transfer fallacious social intentions (Kaitz et al., 2004). If individuals with WS are more likely to approach strangers, invade their personal space and not have an awareness of the dangers this could pose, then they could be facing significant levels of risk during social interactions (Lough et al., 2015a). These issues become even more problematic when combined with the reduced intellectual functioning of individuals with WS (Searcy et al., 2004), staring at faces (Riby & Hancock, 2008), and problems interpreting socio-communicative signals (Porter et al., 2007) meaning that they may miss important subtle cues from those with whom they are approaching and interacting.

The role of the neural structures underlying social behaviour in WS is of particular interest, given the current findings on atypical personal space regulation. Frontal lobe dysfunction has been related to the hypersociability behavioural phenotype observed in WS. It is known that regions in the frontal lobe are involved in regulating and suppressing socially inappropriate actions (Meyer-Lindenberg et al., 2005). Porter et al. (2007) drew parallels between social approach behaviour in individuals with WS, and individuals with frontal lobe

damage. They observed how both groups displayed impulsive social behaviour, which they attributed to impairments in response inhibition. Indeed, Little et al. (2013) proposed that frontal-lobe controlled response inhibition was indicative of social approach behaviour. Frontal lobe theory and lack of inhibitory control have also been implicating in ASD (Christ et al. 2007), leading Lough et al. (2015a) to suggest that difficulty with inhibitory control could be used to explain atypical social distancing regulation in individuals with WS and with ASD.

Alternatively, the amygdala theory has been proposed to help explain the atypical social behaviour seen in WS, and indeed ASD (Jawaid et al., 2012). The amygdala is involved in processing and recognising emotions from faces, generating and controlling anxiety, and mediating eye gaze (Fried, MacDonald & Wilson, 1997). Kennedy et al. (2009) showed that a patient with bilateral amygdala damage showed considerably reduced personal space boundaries. Increased amygdala volume has been repeatedly found in individuals with WS (e.g. Haas et al., 2014). When viewing faces, individuals with WS show reduced activation of the amygdala compared to controls (Kliemann et al. 2012). It may therefore be that abnormalities in amygdala development could be central to the deficits in social judgement and face perception processing seen in WS, which results in atypical emotional reactions and social behaviour linked to social distance regulation. However, as emphasised by Lough et al. (2015a), the current methodology does not allow for further differentiation between these explanations due to the methodology used.

There was no significant relation between age and how close young people with WS stood from other people, but there was a relation between age and how close they let other people stand from them (although caution is advised when interpreting these findings due to the small sample size). Older individuals asked the unfamiliar person to maintain a greater distance than the younger participants did, implying that as they get older, they become more

protective of their own personal space boundaries, but still lack awareness of invading other people's personal space boundaries. It is thought that children display adult-like personal space regulation by age 12 years (Aiello, 1987), which could explain why there were no relation between age and personal space in the TD group who had a mean age of 11.3 years.

The relationship between interpersonal distance and age is less clear when looking at the findings from the SRS. Results from the current study show that there was a significant negative correlation between age and Item 55 ("Knows when he or she is too close to someone, or is invading their personal space"), suggesting that as children get older, their parents rate them as being less likely to invade the personal space of other people. This is in contrast to both the behavioural data collected from the stop-distance task, and also the findings from Lough et al. (2015a), who did not observe any age related changes in interpersonal distance for individuals with WS. However, Lough et al. (2015a) employed a wide age range of 4 – 36 years and, for the purposes of their age analyses, split the sample in to "childhood", "adolescence" and "adulthood" age categories, which could not be done in the present study. It seems likely that the different age range of the sample and analysis method contributed to the differences observed. Future work tracking the developmental trajectory of personal space regulation in individuals with WS, and other developmental disorders, is warranted.

Interestingly, our results show that there was not a significant correlation between SRS scores addressing personal space and distance on the stop-distance paradigm, which could explain why the relationship between age and interpersonal distance differed depending on the methodology used. As the items on the SRS relating to personal space are scored on a 0-3 scale, it is likely that there is not enough variability in scores to allow for meaningful correlations (in addition to a relatively small sample size). Nevertheless, the reliability of

measures used to assess personal space still requires further investigation, particularly in individuals with an intellectual disability (ID).

Despite the contrasting findings on the impact of age, there are congruent findings of atypical personal space regulation in WS across studies (e.g. Lough et al., 2015a, Gessaroli et al., 2013, Kennedy & Adolphs, 2010) and methodologies (i.e., SRS and stop-distance paradigm). Yet, findings on personal space in ASD show discrepant findings depending on the methodology used. Like individuals with WS, individuals with ASD experience high levels of anxiety (Rodgers et al., 2012). Recent work by Perry et al. (2015) suggested that the discrepancy in findings on interpersonal distance in the ASD literature could be partially explained by levels of social anxiety (SA). Perry et al., (2013) found a positive correlation between SA traits and interpersonal distance preference, with individuals with high SA traits preferring to stay further away from a stranger compared to those with low SA traits. Future work on interpersonal distance in ASD would therefore benefit from careful consideration of the anxiety profiles of the participants included in the sample.

This study offered a novel insight into the issue of personal space regulation in young people with WS, and has clear real world implications for these individuals. However, the limitations of this work merit attention. First, a relatively small sample size was used. This is due to the rarity of the condition, meaning relatively small sample sizes are often seen across the WS literature. The number of participants is also comparable to the sample size of the previously mentioned stop-distance study by Gessaroli et al. (2013) involving children with autism. Second, the stop-distance paradigm could be seen as an artificial task, and may not capture behaviour that is reflective of real life. This task has been used in several previous studies on personal space (e.g. Gessaroli et al., 2013; Kennedy et al., 2009), but further observational work would be welcomed to offer more insight into this behaviour. Third, while the findings appear congruent with the WS behavioural phenotype, it is unclear

whether the patterns observed are specific to WS, or rather a feature of having an ID. Future work would therefore benefit from the inclusion of an ID comparison group. Fourth, IQ data were not collected for participants in the current study. Previous work with individuals with ASD has shown that social distancing abnormalities cannot entirely be explained by intelligence levels (e.g. Kennedy & Adolphs, 2014); however, the inclusion of IQ assessment is an important next step in furthering the work on interpersonal distancing in WS. This would be particularly useful in further exploring the age-related findings outlined in the current study. Finally, although the familiarity of the person conducting the stop-distance task was manipulated, all testing took place in familiar environments. It may be, therefore, that the young people with WS viewed the stranger as a ‘trusted stranger’. Whilst this would be true for both the WS and the TD groups, it is likely that individuals with WS have more experience of unfamiliar professionals interacting with them which could impact upon their behaviour. This further emphasises the need for future work to include an ID comparison group, in order to draw conclusions on syndrome-specific patterns of personal space regulation.

In conclusion, the current study offers a new insight into social distance regulation in young people with WS. These issues become especially concerning in light of the constellation of issues/abilities we associate with the disorder – it is when you look at them all together that the vulnerabilities are so enhanced. When considering the wider social profile associated with WS, these findings feed into what is already known about social vulnerability in these individuals (Jawaid et al., 2012), and raise significant concerns about their safety when interacting with strangers. Future work needs to expand on this with large sample, cross-syndrome studies, in order to gain more insight into the degree of disorder-specific patterns of personal space regulation. It is also important to consider the developmental trajectory associated with personal space regulation, and the real world implications

associated with this. Adults with WS are more likely to be independent and have more encounters with strangers, and as a result could face a greater degree of risk by displaying atypical social distancing.

Acknowledgements

The authors acknowledge the work of Eleanor Kerry and Lydia Barge towards data collection for the TD sample. The authors also acknowledge Daniel Kennedy for his advice and guidance in early stages of study preparation. The work would not have been possible without the support of the Williams syndrome Foundation UK and all families who participated.

References

- Aiello, J. R. (1987). Humans spatial behaviour, In D. Stokols & I. Altman (Eds.). *Handbook of environmental psychology* (pp. 389 – 504). York: Wiley
- Constantinno, J., Gruber, C. (2005). The social responsiveness scale: Western Psychological Services
- Christ, S. E., Holt, D. D., White, D. A., & Green, L. (2007). Inhibitory control in children with autism spectrum disorder. *Journal of Autism and Developmental Disorders*, 37(6), 1155–1165
- Fried, I., MacDonald, K.A. & Wilson, C.L. (1997). Single neuron activity in human hippocampus and amygdala during recognition of faces and objects. *Neuron* 18, 753–765
- Gessaroli, E., Santelli, E., di Pellegrino, G., Frassinetti, F. (2013). Personal space regulation in childhood autism spectrum disorders. *PLoS One*, 8 (9), e74959
- Greenberg, C. I., Strube, M. J., & Myers, R. A. (1980). A multitrait-multimethod investigation of interpersonal distance. *Journal of Nonverbal Behavior*, 5(2), 104-114.
- Hall, E. (1966). *The Hidden Dimension*. Garden City, New York: Doubleday
- Haas, B.W., Sheau, K., Kelley, R.G., Thompson, P.M. & Reiss, A.L. (2014). Regionally specific increased volume of the amygdala in Williams syndrome: Evidence from surface-based modeling. *Hum. Brain Mapp.* 35, 866–874
- Hayduk, L. A. (1978). Personal space: An evaluative and orienting overview. *Psychological bulletin*, 85(1), 117.
- Hayduk, L. A. (1983). Personal space: Where we now stand. *Psychological bulletin*, 94(2), 293.
- Hayduk, L. A. (1985). Personal space: The conceptual and measurement implications of structural equation models. *Canadian Journal of Behavioural Science/Revue canadienne des sciences du comportement*, 17(2), 140

- Hillier, L., Fulton, R., Fulton, L., Graves, T., Pepin, K., Wagner-McPherson, C. et al. (2003). The DNA sequence of human chromosome. *Nature*, 7 (424), 157 - 164
- Järvinen A., Korenberg J. R., Bellugi U. (2013). The social phenotype of Williams syndrome. *Current Opinion in Neurobiology*. 23, 414–422
- Jawaid, A., Riby, D., Owens, J., White, S., Tarar, T., & Schulz, P. (2012). ‘Too withdrawn’ or ‘too friendly’: considering social vulnerability in two neuro-developmental disorders. *Journal of Intellectual Disability Research*, 56(4), 335–350.
- Jones, W., Bellugi, U., Lai, Z., Chiles, M., Reilly, J., Lincoln, A., & Adolphs, R. (2000). Hypersociability in Williams syndrome. *Journal of Cognitive Neuroscience*, 12, 30-46.
- Kaitz, M., Bar-Haim, Y., et al., (2004). Adult attachment style and interpersonal distance. *Attachment and Human Development*. 6 (3), 285–304
- Kennedy, D.P. & Glascher, J., Tysza, J. & Adolphs, R. (2009). Personal Space Regulation by the human amygdala. *Nature Neuroscience*, 12, 1226 – 1227
- Kennedy, D.P. & Adolphs, R. (2014). Violations of personal space by individuals with autism spectrum disorder. *PLoS One*, 9 (8), e103369
- Klein-Tasman, B.P., Li-Barber, K.T., Magargee, E.T. (2011). Honing in on the social phenotype in Williams syndrome using multiple measures and multiple raters. *Journal of Autism and Developmental Disabilities*, 41(3), 341-351.
- Kliemann, D., Dziobek, I., Hatri, A., Baudewig, J. & Heekeren, H.R. (2012). The role of the amygdala in atypical gaze on emotional faces in autism spectrum disorders. *J. Neurosci.* 32, 9469–9476
- Korenberg J. R., Bellugi U., Salandanan L. S., Mills D. L. & Reiss A. L. (2003) Williams Syndrome: aneurogenetic model of human behaviour. In: *Nature Encyclopaedia of the Human Genome*, pp. 757–66. Nature Publishing Group, London, UK

- Little, K., Riby D. M., Janes, E., Clark, F., Fleck, R. & Rodgers, J. (2013). Hetrogeneity of social approach behaviour in Williams syndrome: The role of response inhibition. *Research in Developmental Disabilities*, 34, 959 – 967
- Lough, E., Flynn, E. & Riby, D. M. (2015b). Mapping real-world to online vulnerability in young people with developmental disorders: Illustrations from autism and Williams syndrome. *Review Journal of Autism and Developmental Disorders* 2(1), 1-7.
- Lough, E., Hanley, M., Rodgers, J., South, M., Kirk, H., Kennedy, D. P., & Riby, D. M. (2015a). Violations of Personal Space in Young People with Autism Spectrum Disorders and Williams Syndrome: Insights from the Social Responsiveness Scale. *Journal of autism and developmental disorders*, 1-8.
- Meyer-Lindenberg, A. Hariri, A., Munoz K., Mervis, C., Mattay, V., Morris, C. et al. (2005) Neural correlates of genetically abnormal social cognition in Williams syndrome. *Nature Neuroscience*, 8, 991–993
- Nauta, M. H., Scholing, A., Rapee, R. M., Abbott, M., Spence, S. H., & Waters, A. (2004). A parent-report measure of children's anxiety: psychometric properties and comparison with child-report in a clinic and normal sample. *Behaviour research and therapy*, 42(7), 813-839.
- Perry, A., Levy-Gigi, E., Richter-Levin, G., & Shamay-Tsoory, S. G. (2015). Interpersonal distance and social anxiety in autistic spectrum disorders: a behavioral and ERP study. *Social neuroscience*, 10(4), 354-365
- Perry, A., Rubinsten, O., Peled, L., & Shamay-Tsoory, S. G. (2013). Don't stand so close to me: A behavioral and ERP study of preferred interpersonal distance. *Neuroimage*, 83, 761-769
- Porter, M. A., Coltheart, M., & Langdon, R. (2007). The neuropsychological basis of hypersociability in Williams and Down syndrome. *Neuropsychologia*, 45(12), 2839–2849.
- Riby, D., Hanley, M., Kirk, H., Clark, F., Little, K., Fleck, R. et al. (2014). The Interplay Between Anxiety and Social Functioning in Williams Syndrome. *J Autism Dev Disord.*, 13, 1984-1987

- Rizzolatti, G., Fadiga, L., Fogassi, L., & Gallese, V. (1997). The space around us. *Science*, 277(5323), 190.
- Rodgers, J., Riby, D. M., Janes, E., Connolly, B., & McConachie, H. (2012). Anxiety and repetitive behaviours in autism spectrum disorders and Williams syndrome: A cross syndrome comparison. *Journal of Autism and Developmental Disorders*, 42, 175–180
- Sambo, C. F., Liang, M., Cruccu, G., & Iannetti, G. D. (2012). Defensive peripersonal space: the blink reflex evoked by hand stimulation is increased when the hand is near the face. *Journal of neurophysiology*, 107(3), 880-889
- Searcy, Y.M., Lincoln, A., Rose, F., Klima, E., Bavar, N., & Korenberg, J.R. (2004). The relationship between age and IQ in adults with Williams syndrome. *American Journal on Mental Retardation*, 109, 231–236
- Sparaci, L., Stefanini, S., D'Elia, L., Vicari, S., & Rizzolatti, G. (2014). What and why understanding in autism spectrum disorders and Williams syndrome: Similarities and differences. *Autism Research*, 7(4), 421-432.
- Sparaci, L., Stefanini, S., Marotta, L., Vicari, S., & Rizzolatti, G. (2012). Understanding motor acts and motor intentions in Williams syndrome. *Neuropsychologia*, 50(7), 1639-1649
- Tager-Flusberg, H., & Sullivan, K. (2000). A componential view of theory of mind: Evidence from Williams syndrome. *Cognition*, 76(1), 59-90
- Vignemont, F., & Iannetti, G. D. (2015). How many peripersonal spaces? *Neuropsychologia*, 70, 327-334.

Figure Caption Sheet

Figure 1. Stop-distance paradigm. In condition A, the participant approached the experimenter (unfamiliar). In condition B, the experimenter (unfamiliar) approached the participant. In condition C, the participant approached their parent (familiar). In condition D, the parent (familiar) approached the participant.

Figure 2. The percentage of scores on item 52, item 55, item56 and item 63 on the SRS for children with WS and TD children. Higher scores indicate greater atypicalities of behaviour.

Figure 3. Interaction graphs showing interpersonal distance when the child is being approached by / is approaching familiar and unfamiliar people.

Figure 1 top

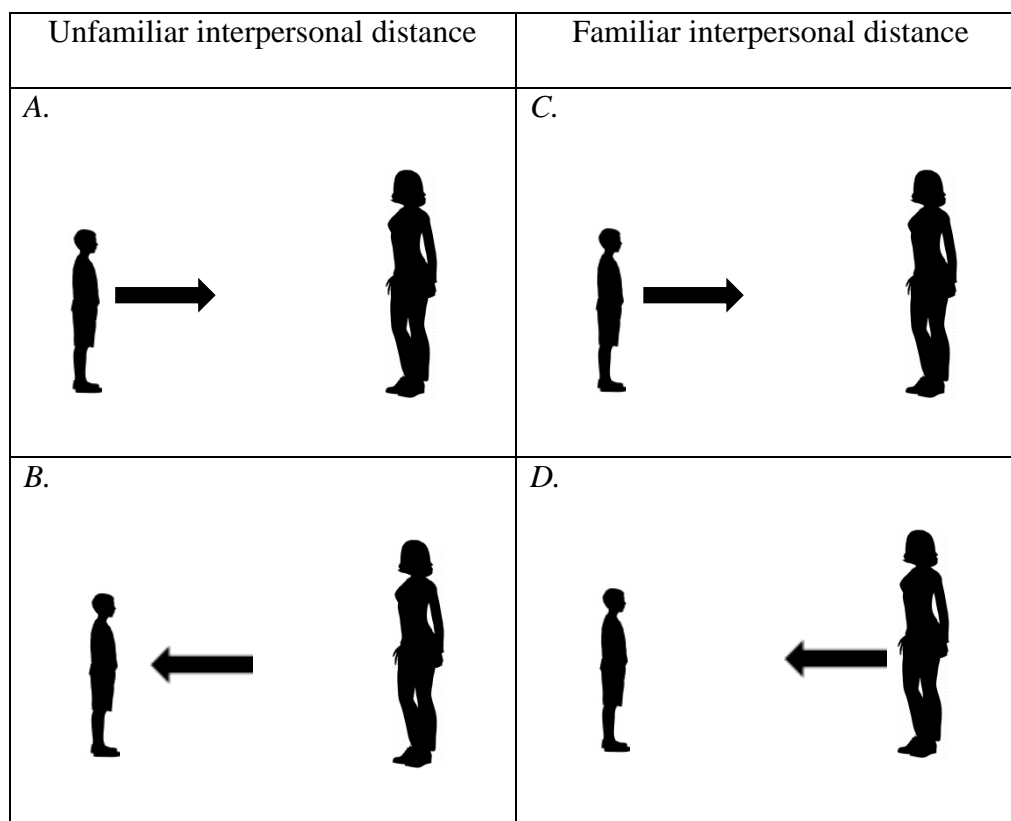
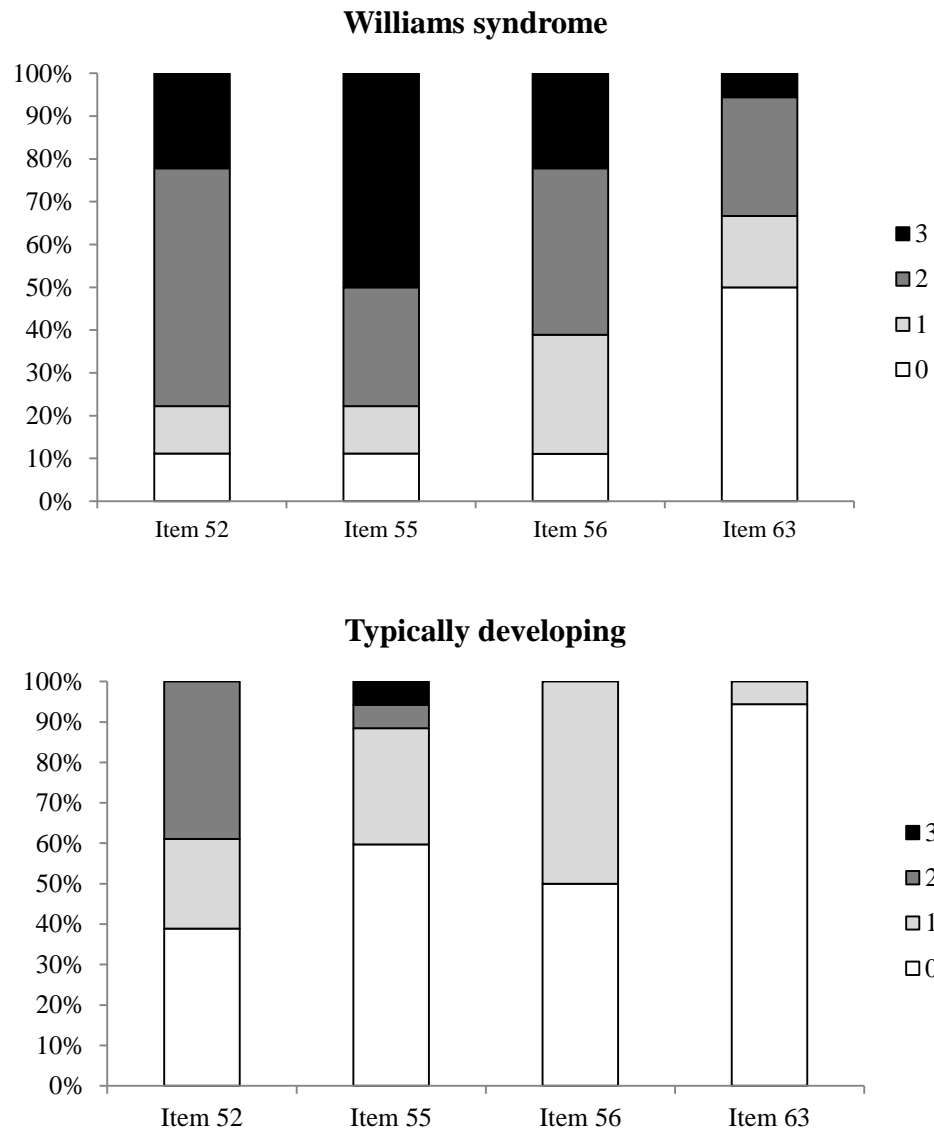


Figure 2 top



* Item 52: “Knows when he or she is talking too loud or making too much noise; item 55: “Knows when he or she is too close to someone or is invading someone’s space”; item 56: “Walks in between two people who are talking”; item 63: “Touches others in an unusual way e.g. he or she may touch someone just to make contact with them then walk away without saying anything”.

Figure 3 top

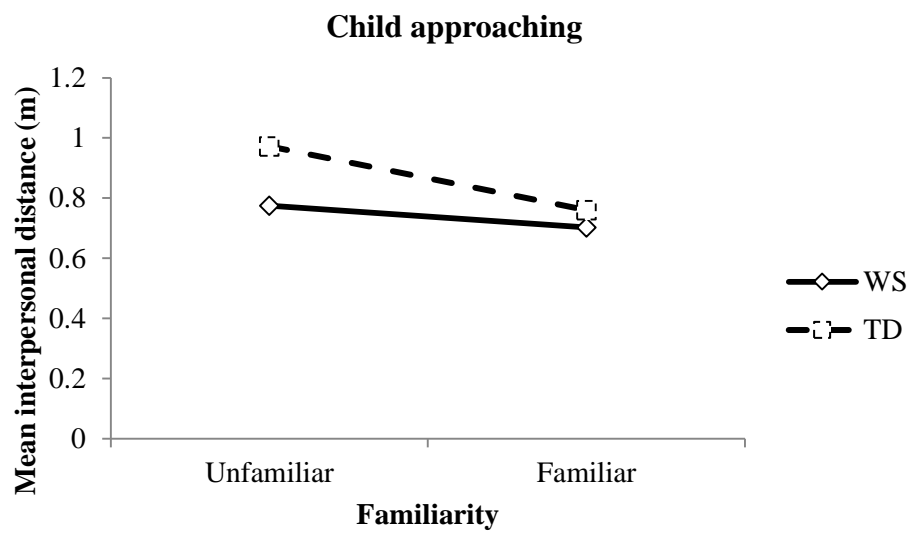
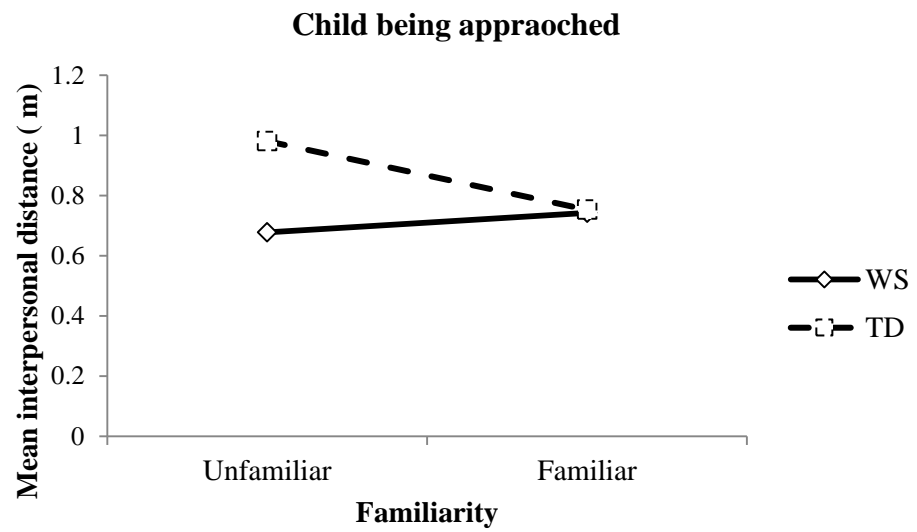


Table 1. Participant characteristics and SRS scores for individuals with WS and those who are typically developing

	WS (n = 18)	TD (n = 18)	<i>p</i> value
Mean age (\pm SD)	11.4 (\pm 2.5) years	11.3 (\pm 2.5) years	
Males/Females (%)	44/56	44/56	
<i>SRS T scores</i>			
Total score	77.17 (\pm 13.37)	44 (\pm 7.98)	*
Social awareness	66.06 (\pm 11.53)	45.67 (\pm 8.04)	*
Social cognition	79.72 (\pm 10.34)	44.22 (\pm 6.62)	*
Social communication	74.5 (\pm 12.95)	46.83 (\pm 8.72)	*
Social motivation	63.39 (\pm 12.57)	47.5 (\pm 7.08)	*
Autistic mannerisms	82.22 (\pm 12.25)	46.28 (\pm 6.09)	*

* = $p < 0.001$